CASE REPORT

Castleman disease of the neck: A case report

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Abstract

Castleman disease is a rare lymphoproliferative disorder with two distinctive presentations, namely Unicentric disease and Multicentric disease. Unicentric disease occurs as a solitary enlarged mass in the mediastinum and is rarely found in the head and the neck, whereas, Multicentric disease appears as a systemic disease with peripheral lymphadenopathy. Here we discuss an unusual case with characteristic clinical, radiologic and histologic findings of a 22-year-old male with Unicentric disease demonstrating a slow growing neck mass often times misdiagnosed as a lymphoma.

Keywords: Castleman disease, Lymph nodes, Biopsy.

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Introduction

In 1956, Dr. Benjamin Castleman first described a series of patients with solitary hyperplastic mediastinal lymph node.[1] Cases initially reported, had a localized disease process with minimal systemic symptoms. However, later on a systemic form of the disease was described, which was characterized by multiple symptoms which included recurrent fevers, night sweats, hepatosplenomegaly and generalized lymphadenopathy. These two forms of the disease are now known as Unicentric disease (UCD) and Multicentric disease (MCD), respectively. The pathogenesis of Castleman disease (CD) is poorly understood, although some studies have suggested the overexpression of Interleukin-6 from the affected lymph nodes.[2,3]

The most common presentation of CD includes solitary enlarged mediastinal lymph nodes (70% of cases), however, the involvement of other locations like pelvis, abdomen and retroperitoneum have also been reported.[4] Patients complain of cough and dyspnoea while the radiographic investigations reveal a 5-9 cm painless mediastinal mass. Here, we present an unusual case of UCD with a neck mass. This case highlights some of the distinctive features of CD and serves as an instructive example for the workup, diagnosis and management of the disease.

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Case Report

The case was first seen in January, 2014 in ENT clinic at Aga Khan University Hospital, Karachi, Pakistan. A 22-year-old male presented with a large and painless mass on the left side of the neck which had been increasing in size gradually for the past 8 years. He otherwise had no symptoms and denied any history of fever, cough, dyspnoea, weight loss, fatigue or night sweats. Patient did not have any co-morbid conditions as asthma, hypertension, diabetes or tuberculosis. He denied the use of alcohol, recreational drugs or smoking but was addicted to oral chewable tobacco locally found in the market. An ultrasound guided fine needle aspiration cytology (FNAC) of the mass revealed a reactive lymph node. Patient was then prescribed a broad spectrum antibiotic (Intra Venous Ceftriaxone 2g once daily) for 10 days but showed no decrease in the size of the mass.

On clinical examination, it was a soft, mobile and a non-tender mass approximately 5 x 5 cm in size on the left side of the neck at level V without any overlying skin changes or other lymph node enlargements. Fiber-optic nasopharyngoscopy showed small amounts of adenoid

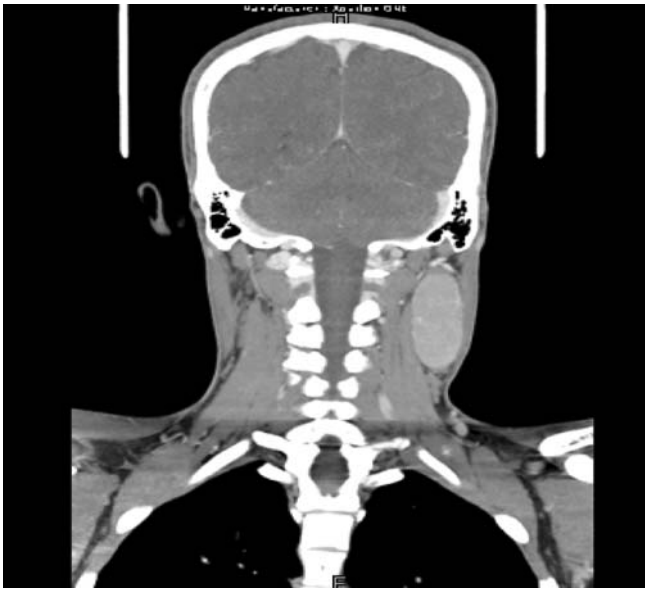

Figure-1: CT scan of the head and neck demonstrated a lobulated enhancing soft tissue mass in the left sided posterior triangle of the neck measuring approximately 56 x 37 x 25 mm in maximum craniocaudal AP and transverse dimensions.

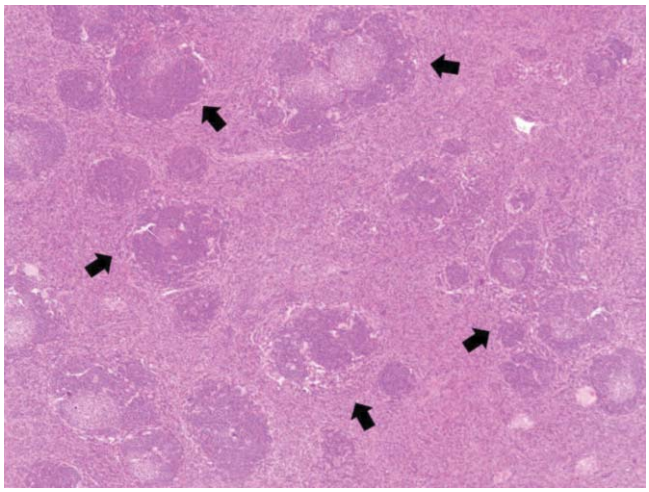

Figure-2: Lymph node with altered architecture exhibiting regressed lymphoid follicles in different stages (arrow heads). Lack of sinuses is also appreciated. (H&E stain, 40x magnification).

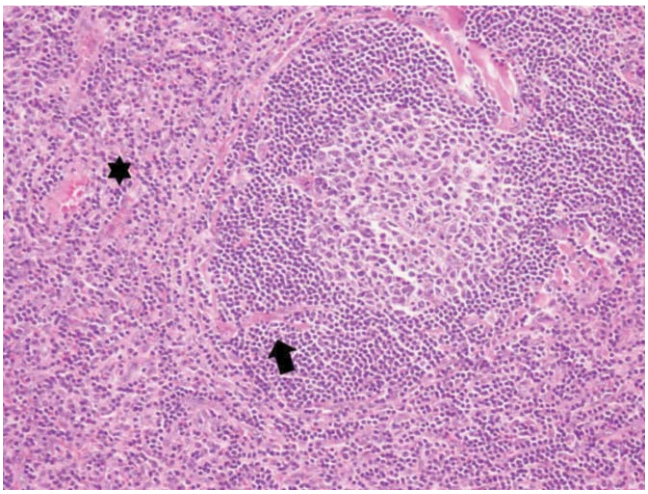

Figure-4: Interfollicular areas showing proliferation of post-capillary endothelial venules (star). A hyalinized blood vessel is traversing the lymph node (lollipop sign) is also appreciated (arrow head). (H&E stain, 200x magnification).

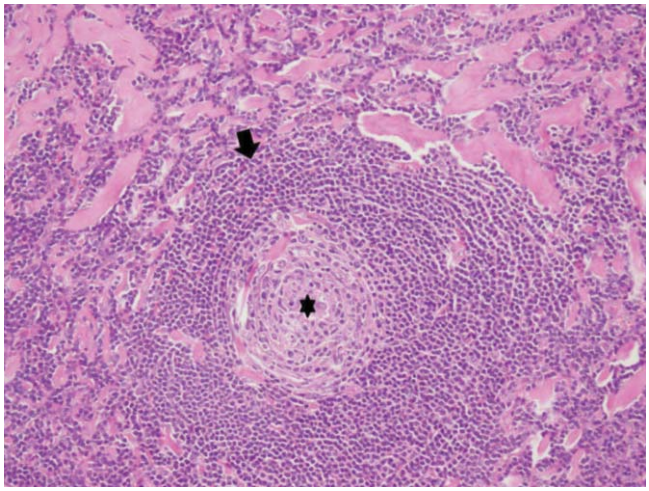

Figure-3: Regressed germinal center comprising only of follicular dendritic cells along with focal hyalinization within germinal center (star). The lymphocytes in the expanded mantle zone are arranged in concentric layers/onion-skin pattern (arrow head). (H&E stain, 200x magnification).

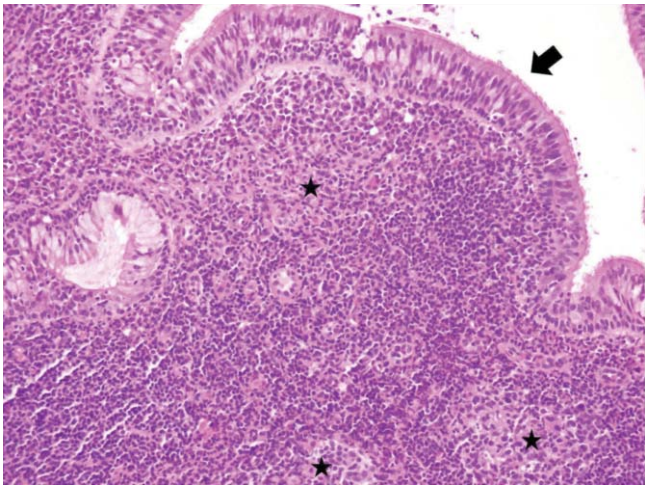

Figure-5: Nasopharynx biopsy showing respiratory mucosa (arrow head). Dense lymphoid infiltrate is appreciated in subepithelial tissue along with lymphoid follicles showing germinal centers (stars). (H&E stain, 200x magnification).

tissue occupying the nasopharynx. Computed Tomography (CT) scan of the head and the neck demonstrated an enhanced lobulated soft tissue mass on the left sided posterior triangle of the neck, measuring approximately 56 x 37 x 25 mm in the maximum cranio-caudal, anterio-posterior (AP) and transverse dimensions, respectively. This suggests a possibility of a neurogenic lesion with a less likely chance of an enlarged solitary lymph node (Figure-1). A few centimeters away, some cervical lymph nodes were identified involving levels I, II, III and V, which were likely to be reactive in nature.

Subsequently, the patient underwent an excisional biopsy of the lymph node and the adenoid tissue in nasopharynx. Intra-operative findings included, a left sided neck mass approximately 4 x 4 cm in size involving the levels II and level III of the neck. Entire lymph node was removed with an intact capsule and sent for histopathology. Small adenoid tissues removed by trans-nasal endoscopic approach were also sent for histopathology.

On microscopic histopathological examination, lymph nodes revealed an altered architecture exhibiting loss of sinuses and lymphoid follicles in various stages of regression (Figure-2). The regressed germinal centers were devoid of lymphocytes and showed residual follicular dendritic reticulum. The mantle zone was expanded by

mature lymphocytes which were arranged in concentric (onion-skin) fashion (Figure-3). The interfollicular regions were expanded and showed proliferation of post-capillary endothelial venules and hyalinization. A mixed population of lymphocytes and plasma cells were seen in the interfollicular regions. Occasional venules traversing the lymphoid follicles were also visible (Figure-4).

The tissue from the nasopharynx revealed a benign adenoid tissue exhibiting lymphoid follicles with prominent germinal centers. (Figure-5).

Patient was followed one year after surgery and was found to be well.

Discussion

Since its first recognition in 1956, understanding of Castleman disease has become more evident after the segregation of two entities namely, Unicentric and Multicentric disease forms showing distinctive prognosis and treatment approaches. We present an unusual case of UCD with a nasopharyngeal mass with lymph node involvement in the neck.

The incidence and prevalence of CD has become difficult to evaluate due to its rare occurrence in the general population, however, it is seen to affect both the sexes equally.[4] UCD is usually found to have a peak incidence in the second to fourth decades of life, while MCD tends to present later in the sixth and seventh decades.[4] In majority of the cases, UCD occurs as an isolated mediastinal lymph node enlargement with few patients complaining of cough, dyspnoea or other constitutional symptoms. MCD on the other hand, has a constellation of systemic symptoms with peripheral lymphadenopathy involving multiple compartments throughout the neck, chest, abdomen and pelvis. It also has an association with human immunodeficiency virus (HIV) and human herpesvirus-8 (HHV-8) infections.[5]

The differential diagnosis of UCD includes lymphoma and any other causes of lymphadenopathy. Initial investigations encompasses a CT scan which reveals the location, extent and number of lymph nodes involved which are then visualized with a homogenously intense contrast enhancement.[6] Magnetic Resonance Imaging (MRI) can be used to further clarify the involvement of any soft tissues. Obtaining a cytologic or tissue diagnosis is preferentially the next step. FNAC cannot reliably predict CD but helps to rule out metastatic carcinoma and most often Non-Hodgkin's lymphoma. Excisional biopsy is generally required to establish the final diagnosis.

Surgical excision has proven to be curative for UCD. In 16 cases reported by Bowne et al.[7] no recurrence was found either clinically or radiologically in the 8 cases that underwent surgical excision. Similar results supporting the surgical excision have been reported by Herrada et al.[8] and Chronowski et al.[9] Moreover, surgical excision was seen to decrease the mortality rates (17.6% to 3.8%) in UCD in a systematic review of 404 published cases.[10] In contrast, MCD had a poor response to surgery and worse prognosis when compared to UCD. MCD does not have a proven treatment regimen, however, chemotherapy with rituximab has been tried earlier with varying results.

Conclusion

CD mimics lymphoma in its presentation and is often misdiagnosed. Fine needle aspiration usually turns out to be of little value and does not yield a definitive diagnosis which makes excisional biopsy an imperative choice. Physicians, Otolaryngologists and Pathologists should be vigilant while managing painless expanding neck mass and consider a possibility of CD. Correct diagnosis will hasten the treatment strategy especially, in cases of Unicentric variety where surgical excision can be curative in nature.

Disclaimer: None to declare.

Conflict of Interest: None to declare.

Funding Sources: None to declare.

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